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Focal Fibrocartilaginous Dysplasia of Long Bones: Report of Eight Additional Cases and Literature Review

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Summary: We report eight additional cases of focal fibrocartilaginous dysplasia (FFCD) in the proximal tibia (five), distal ulna (one), and distal femur (two). Spontaneous, complete resolution of the lesion was observed in two tibiae and one ulna. Three tibial lesions with genu varum deformity were managed with osteotomy. Two femoral FFCDs caused persistent or progressive deformity: one genu valgum with patellar dislocation, and one genu varum. These patients underwent concomitant deformity correction and lengthening by the Ilizarov method. The final results were satisfactory in all patients except one, who underwent valgus tibial osteotomy and developed mild postoperative genu valgum. The analysis of a total of 46 cases in the literature and our experience suggests that (a) FFCD has a wide histopathologic spectrum, ranging from purely dense, fibrous tendon-like tissue to benign fibrocartilaginous tissue; (b) at least 45% of tibial FFCD demonstrates progressive, spontaneous resolution; (c) in contrast, femoral and humeral FFCDs appear to have a slim possibility of spontaneous regression of the deformity; and (d) corrective osteotomy is indicated when the deformity is increasing or persistent, or when the existing deformity is severe enough to jeopardize adjacent joint mechanics and alignment. **Key Words:** Focal fibrocartilaginous dysplasia—Ilizarov—Osteotomy.

Focal fibrocartilaginous dysplasia (FFCD) is an uncommon, benign condition that has been known to cause deformity of the long bone in young children. A total of 38 cases (33 tibia, three femur, one humerus, one ulna) have been reported in the literature (Table 1). Bell et al. (3) postulated that the mesenchymal anlage in the area of the pes anserinus, which fails to differentiate normally and persists as a focus of fibrocartilage, could interfere with the growth of the medial aspect of the proximal tibia to produce varus deformity. Previously reported FFCD of the femur and upper extremity (1,14) were also composed of dense fibrous tissue with areas of cartilaginous differentiation, but without an abnormal tendon insertion. The purpose of this study was to report eight additional cases of FFCD of the long bones and to combine our data with a compilation of the cases from the literature. We sought to develop a better understanding of the histopathologic spectrum, the evolution of the lesion according to the affected site, and responses to conservative and operative treatments.

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MATERIALS AND METHODS

Between February 1993 and May 1998, we treated eight additional cases of FFCD of the long bones in children: five in the proximal tibia, two in the distal femur, and one in the distal ulna. In addition, data about 38 children with FFCD of the long bones were compiled from 15 previously published reports (12 in English, two in French, and one in Italian) identified through a Medline search. The data collected included age at presentation, gender, affected anatomic site, presenting deformity, resolution or progression of deformity, histopathologic findings, treatment methods, outcome, complications, and residual problems.

CASE REPORTS

Case 1

A 17-month-old boy was first seen for an anterolateral bowing of the right thigh, which had been noticed at the age of 11 months by his parents. On examination, there was also a 20° medial torsion of the lower leg, and 1 cm of leg-length shortening. Plain radiographs revealed an ill-defined cortical lesion of posteromedial aspect of the distal femur, associated with a 26° varus tibiofemoral angle on anteroposterior projection (Fig. 1A). At the age of 33 months, this angle increased to 42° (Fig. 1B). Mag-

TABLE 1. *Details of 46 patients with FFCD*

Author (yr)	Age at present (mo)	Sex, side, location	Deformity (°)		Shortening (mm)	Deformity	Management	Age at operation (mo)	Age at final follow-up (mo)	Histologic findings			Final outcome, complication
			Varus	Internal torsion						FB	FC	HC	
Bell	9	M,Rt,PT	20	30	10	Pro	Osteotomy	24	78	+	+	–	NA
(1985)	15	M,Rt,PT	+	30	–	Pro	Osteotomy	24	36	+	+	–	NA
	27	F,Rt,PT	+	?	5	Reg	Observation		84				NA
Bradish	15	F,Lt,PT	?	?	?	?	Osteotomy	28	?	+	+	–	?
(1988)	24	M,Rt,PT	?	?	?	Reg	Observation		48				NA
	17	M,Rt,PT	?	?	?	Reg	Biopsy	28	55	+	+	–	NA
	28	F,Lt,PT	?	?	?	Reg	Observation		43				RV
	16	F,Rt,PT	?	?	?	Per	Osteotomy	25	175	?	?	?	VD, PN
Huisen (1989)	22	M,Rt,PT	+	–	15	Reg	Night splint		46				NA
Herman	18	F,Rt,PT	+	?	–	Reg	Brace		48				NA
(1990)	25	F,Rt,PT	+	?	?	?	Osteotomy	26	36	+	+	–	NA
	16	F,Lt,PT	?	?	?	?	Brace		?				?
Olney	18	F,Lt,PT	+	+	?	Pro	Osteotomy	30	102	+	–	–	NA
(1990)	5	M,Rt,PT	+	?	?	?	Osteotomy	6	42	+	+	–	NA
	6	M,Lt,PT	30	?	?	Pro	Osteotomy	23	41	+	–	–	NA
Kariya	5	F,Rt,PT	+	?	?	Reg	Observation		54				NA
(1991)	12	F,Lt,PT	+	?	?	Reg	Biopsy	18	30	+	+	–	RV
Zayer	12	F,Rt,PT	45	+	15	Reg	Biopsy	16	360	+	+	–	NA
(1992)	12	F,Rt,PT	30	+	30	Reg	Biopsy	43	84	+	+	–	NA
Landreau-Jolivet	17	M,Rt,PT	30 (MDA)	+	8	Reg	Observation		90				NA
(1992)	11	M,Rt,PT	15 (MDA)	?	?	Pro	Osteotomy	36	96	?	?	?	NA
	11	M, PT	?	?	?	Reg	Observation		?				NA
	13	F, PT	?	?	?	?	Osteotomy		?				NA
Cockshott (1994)	24	M,Lt,PT	+	+	?	?	?		?				NA
Marchiodi (1995)	24	F,Rt,PT	20	+	?	Pro	Osteotomy	?	36	+	+	–	NA
Meyer	7	M,Rt,PT	+	?	?	Per	Osteotomy	26	?	+	–	–	NA
(1995)	12	M,Lt,PT	+	?	?	?	Osteotomy	14	?	+	–	–	NA
Lincoln (1997)	22	M,Rt,PH	ALB	?	+	Pro	Osteotomy	72	120	+	+	–	AD, short 7.7 cm
	24	M,Lt,DU	RVB15	?	+	Reg	Observation		96				RV, short <1 cm
Jouve	11	M,Lt,PT	16	?	10	Reg	Observation		36				RV
(1997)	12	F,Lt,PT	20	+	14	Per	Osteotomy	36	?	+	+	–	?
Albinana (1997)	24	M,Lt,PT	13 (TFA)/ 15 (MDA)	20	10	Reg	Night splint		84				NA
	15	M,Lt,PT	22 (TFA)/ 21 (MDA)	25	10	?	Osteotomy	?	75	+	+	–	NA,PN
	13	M,Lt,PT	10 (TFA)/ 15 (MDA)	10	10	Reg	Night splint		71				RV
	18	M,Lt,PT	17 (TFA)/ 18 (MDA)	10	10	?	Osteotomy	?	65	+	+	–	NA
	16	F,Lt,DF	45 (TFA)	0	15	Pro	Med. periosteal release	?	33	+	+	–	RV
MacNicol	24	M,Lt,DF	25	?	10	Pro	Osteotomy	?	?	+	–	–	NA
(1999)	14	M,Lt,DF	+	?	+	Pro	Osteotomy	?	48	?	?	?	NA
Authors	18	F,Rt,PT	25	20	10	Per	Osteotomy	20	41	+	+	–	NA
(1999)	18	M,Lt,PT	25	20	11	Pro	Periosteal division & incomplete osteotomy	39	59	+	–	–	RV
	16	F,Lt,PT	25	15	10	Per	Osteotomy	33	63	+	+	+	VD
	17	M,Lt,PT	20	–	–	Reg	Observation		75				NA
	17	M,Rt,DF	26 (AB 35)	20	10	Pro	Osteotomy with Ilizarov	33	66	+	+	–	NA

TABLE 1. *(Continued)*

Author (yr)	Age at present (mo)	Sex, side, location	Deformity (°)		Shortening (mm)	Deformity	Management	Age at operation (mo)	Age at final follow-up (mo)	Histologic findings			Final outcome, complication
			Varus	Internal torsion						FB	FC	HC	
	15	F,Lt,DF	Valgus 22 (AB 42)	–	30	Per	Osteotomy with Ilizarov, relocation of patella	25	84	+	+	–	NA
	20	M,Lt,PT	15	–	5	Reg	Observation		32				RV
	25	M,Rt,DU	RB 20	–	15	Reg	Observation		32				RV

Two cases reported by Landreau-Jolivet et al. (13) were not described in detail. They simply mentioned that one case was affected on the right side, and the other, left side; one case showed spontaneous resolution of the lesion, and the other case, which was treated with corrective osteotomy, had normal appearance at the final follow-up.

M, male; F, female; Rt, right; Lt, left; DF, distal femur; DU, distal ulna; PH, proximal humerus; PT, proximal tibia; +, present but not measured; –, absent; ?, no documentation; AB, anterior bowing; ALB, anterolateral bowing; RB, radial bowing; RVB, radiovolar bowing; Per, persistence; Pro, progression; Reg, regression; MDA, metaphyseodiaphyseal angle; TFA, tibiofemoral angle; FB, fibrous tissue; FC, fibrocartilage component; HC, hyaline cartilage island; NA, normal appearance; RV, resolving; VD, valgus deformity; PN, peroneal nerve palsy; AD, acceptable deformity.

netic resonance imaging (MRI) demonstrated fibrous tissue invagination into the medial cortex of the distal femur, which extended proximally (Fig. 1C). Concomitant deformity correction and lengthening was performed at the distal femur using the Ilizarov external fixator. Microscopic examination of the lesion showed both fibrous and fibrocartilaginous elements (Fig. 1D). At the latest follow-up at age 66 months, the lesion was completely healed, and he had a normal, straight leg (Fig. 1E).

Case 2

A 15-month-old girl was seen for genu valgum deformity, which had been noticed at age 12 months. On examination, there was a valgus deformity at the distal femur associated with lateral dislocation of the patella, and 3 cm of leg-length shortening. Radiographs showed a characteristic lesion at the posterolateral aspect of the distal femoral cortex, resulting in 22° of valgus and 42° of anterior bowing. Ten months later, because the deformity persisted without any improvement (Fig. 2A), she underwent concomitant deformity correction and lengthening by the Ilizarov method (Fig. 2B) followed by quadriceps realignment for relocation of the patella. Microscopic examination of the lesion showed dense fibrous tissue invaginating the underlying cortex, admixed with small areas of cartilaginous differentiation, which was confirmed by S-100 immunostaining (Fig. 2C). At the latest follow-up at age 7 years, the lesion had disappeared completely, and she had a normal, straight, and functional leg (Fig. 2D).

Case 3

A 2 year, 1-month-old boy was seen for a deformity of the right forearm, which had been found incidentally without any history of trauma or infection. On examination, there was an obvious 15° radial angulation of the right ulna. Radiographic examination revealed a characteristic focal defect at the distal metaphysis of right ulna, which was shorter than the contralateral normal ulna by 1.5 cm (Fig. 3A). No treatment was given, and the lesion spontaneously resolved at 7 months' follow-up (Fig. 3B).

Case 4

A 17-month-old boy was seen for an obvious deformity of the left leg, which had first been noticed by his parents when he was 3 months old. Radiographic examination revealed a characteristic focal defect consistent with FFCD at the left proximal tibia. There was a 20° varus tibiofemoral angle. No treatment was given, and the patient was lost to follow-up until age 6 years and 3 months. At that time, a complete resolution of the deformity was observed. Radiographs showed spontaneous complete healing of the lesion and a normal, straight leg.

Case 5

An 18-month-old boy had unilateral bowing of the left leg. His parents had noted the deformity when the patient was 6 months old. On examination, there was a 25° varus tibiofemoral angle, a 20° tibial internal torsion, and 1.1 cm of leg-length shortening. Twenty-one months later, the deformity had worsened to a 30° tibiofemoral angle (Fig. 4A and B). Therefore, we decided to explore the lesion. The patient underwent medial hemicircumferential periosteal release and in situ incomplete osteotomy of the proximal tibia at the time of biopsy. Microscopic examination revealed dense collagenous tissue without any cartilaginous element (Fig. 4C). At age 4 years and 11 months, he had an almost normal, straight leg (Fig. 4D).

Case 6

A 12-month-old girl was first seen with bowing of the left leg. The deformity was painless, and there was no history of injury or infection. Examination at age 16 months revealed a 25° varus deformity of the left knee, 15° tibial internal torsion, and 1 cm of leg-length shortening. Radiographs showed a well-defined focal defect of the proximal tibia, highly suggestive of FFCD. At age 33 months, there was a 15° varus deformity and a 15° internal torsion. A proximal tibial valgus derotational osteotomy was performed, and the correction was main-

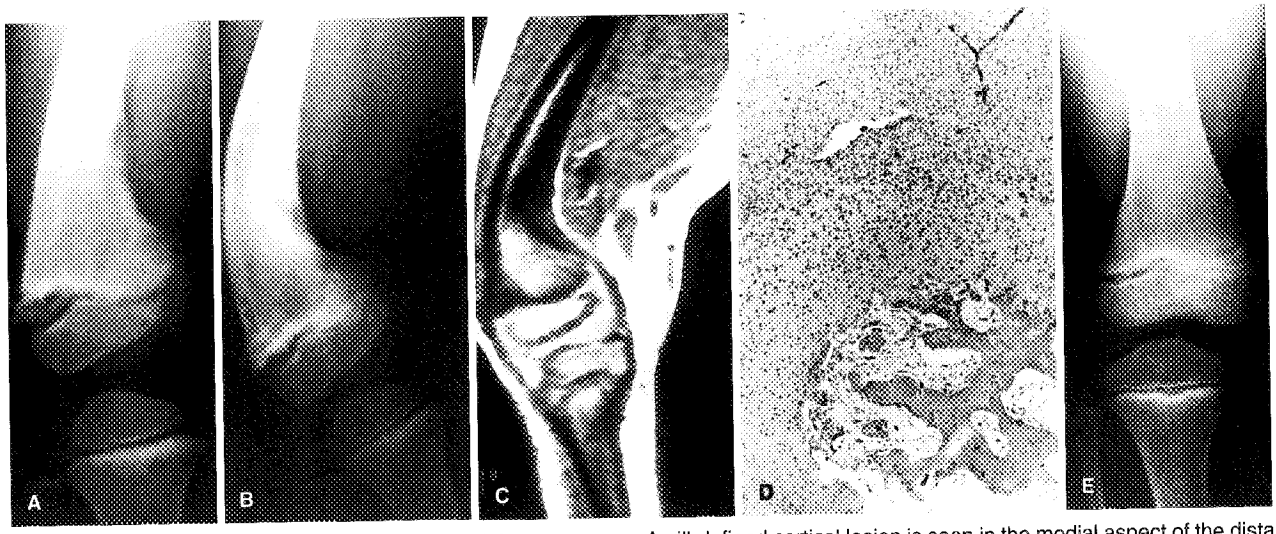

FIG. 1. A: Radiograph in a 17-month-old boy with right genu varum. An ill-defined cortical lesion is seen in the medial aspect of the distal femur. **B:** Radiograph taken at age 33 months shows that the lesion became more discrete, whereas the angular deformity worsened. **C:** MRI (T_1-weighted, TR/TE 390/12) shows intermediate signal intensity of the cortical defect. **D:** Photomicrograph shows that the lesion contains both fibrous (left upper) and fibrocartilaginous (center) elements (H&E, original magnification ×40). **E:** Radiograph taken at 33-month follow-up after concomitant deformity correction and lengthening by the Ilizarov method shows complete resolution of the deformity and excellent alignment.

tained with cross-pinning and plaster. Intraoperatively, the lesion was shown to consist of dense fibrous tissue in the notch on the medial aspect of the tibia into which the pes anserinus was inserted. Microscopic examination demonstrated both hyaline and fibrocartilage with an organoid pattern of transitional zones from fibrous to fibrocartilaginous tissue. Anteroposterior radiographs of the leg, taken immediately postoperatively, revealed a 1° valgus tibiofemoral angle. Two and half years later, although the osteotomy had healed uneventfully, there was a mild valgus deformity of the knee with an 18° tibiofemoral angle, as measured radiographically.

Case 7

An 18-month-old girl had unilateral bowing of the right leg, which had first been noticed by her mother when she began walking at age 12 months. On clinical examination, she had a 25° varus deformity of the right knee, a 20° tibial internal torsion, and 1 cm of leg-length shortening. Radiographs showed a characteristic lesion in the proximal tibia. Two months later, a proximal tibial valgus derotational osteotomy was performed, and the correction was maintained with cross-pinning and plaster. Microscopic examination revealed relatively hypercellular collagenous tissue coupled with well-defined

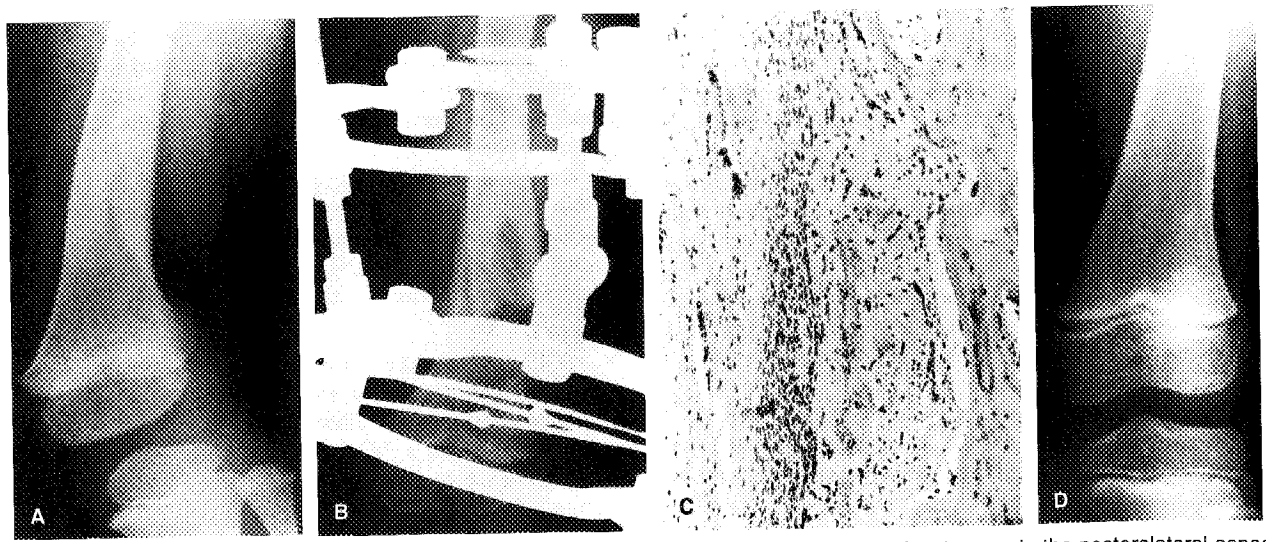

FIG. 2. A: Radiograph in a 25-month-old girl with left genu valgum. A characteristic cortical defect is seen in the posterolateral aspect of the distal femur. **B:** Radiograph shows concomitant deformity correction and lengthening by the Ilizarov method. **C:** Photomicrograph shows dense fibrous tissue invaginating the underlying cortex (right), admixed with small areas of cartilaginous differentiation (H&E, original magnification ×100). **D:** Radiograph taken at age 84 months shows complete resolution of the lesion.

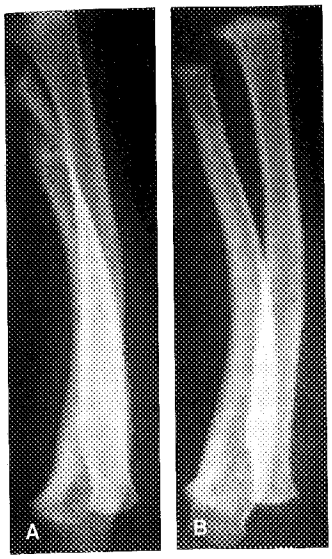

FIG. 3. A: Radiograph in a 25-month-old boy with radial angulation and shortening of the right ulna shows a characteristic focal defect at the distal metaphysis. **B:** Radiograph taken at age 32 months shows complete resolution of the defect.

cartilaginous areas, showing a transition from the fibrous zone. At age 3 years and 5 months, there was complete healing, with a normal, straight leg.

Case 8

A 20-month-old boy was first seen for a 15° genu varum deformity of the left leg. Radiographs revealed a characteristic lesion with a focal defect in the medial aspect of the proximal tibia. No treatment was given, and spontaneous resolution of the deformity was noticed at 1-year follow-up.

RESULTS

Forty-six patients with FFCD have been reported: eight children from the authors' institutions, and 38 from the literature (Table 1). There were 27 boys and 19 girls. Thirty-eight cases of FFCD occurred in the tibia, five in the femur, one in the humerus, and two in the ulna. All had unilateral limb involvement: 22 on the right and 24 on the left. The average age at initial presentation was 16.5 months (range, 5–28 months).

Presenting deformity

All of the patients with tibial FFCD had unilateral varus deformity, usually combined with tibial torsion and sometimes with apparent leg shortening. FFCD occurred in other long bones and resulted in unilateral angular deformities: four cases of distal femoral vara (1,15, and ours), one of distal femoral valga (authors), one of proximal humeral anterolateral angulation (14), and two of distal ulna radial angulation (14, and ours).

Histologic findings

Pathology specimens were obtained during biopsy or corrective osteotomy in 25 cases (Table 1). There were various histologic findings, comprising from dense fibrous tendon-like tissue to fibrocartilaginous tissue and a hyaline cartilage island. Dense fibrous tissue alone without a cartilaginous element was observed in five tibiae (17,18, and ours), and one femur (15). Both fibrous and fibrocartilaginous tissues were seen in 14 tibiae (1,3,4,7,10,11,16,18,19, and ours), three femora (1, and ours), and one humerus (14). One tibial lesion contained both hyaline and fibrocartilages along with fibrous tissue (ours). It was difficult to determine the histopathologic spectrum and evolutionary changes of the lesions from the descriptions of published reports. However, histopathologic studies of our own cases demonstrated that each lesion varied in its histopathologic features. The individual lesions showed regional variation in cellularity and the proportion of fibrous and cartilaginous components. Paucicellular areas were mainly composed of dense fibrous tissue, whereas more cellular areas con-

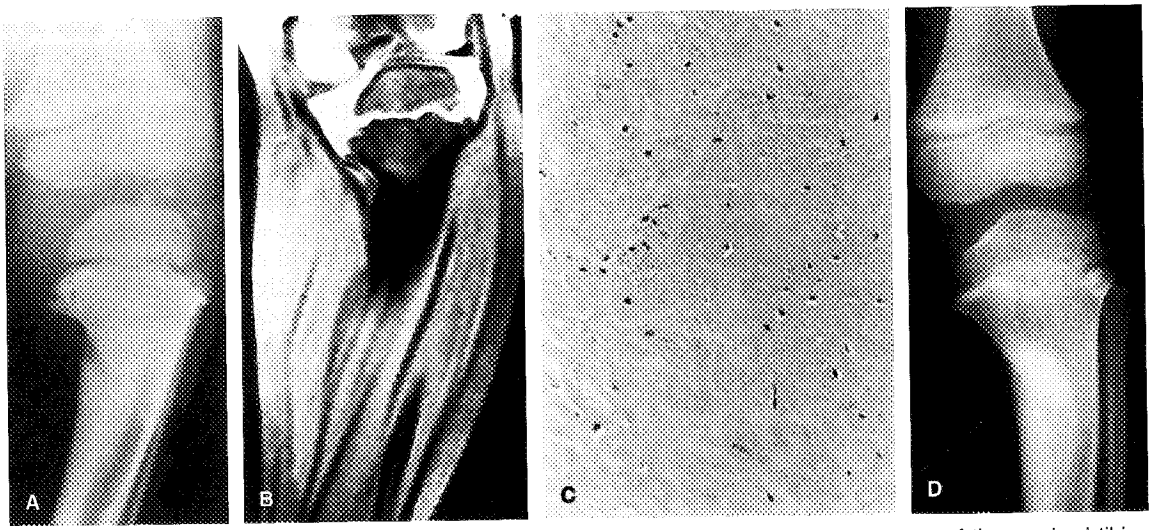

FIG. 4. A: Radiograph in a 39-month-old boy shows a characteristic focal defect in the medial aspect of the proximal tibia. **B:** MRI (gradient echo, TR/TE 600/15, fat suppression) shows low to intermediate signal intensity of the cortical defect. **C:** Photomicrograph shows dense collagenous tissue without any cartilaginous elements (H&E, original magnification ×100). **D:** Radiograph taken at 20 months' follow-up after medial hemicircumferential periosteal release and incomplete osteotomy of the proximal tibia shows complete resolution of the lesion.

tained foci of seemingly metaplastic fibrocartilaginous elements. The cartilage cells in the lesion appeared to undergo degeneration and ossification (enchondral ossification) throughout the remodeling process. Prominent osteoclastic and osteoblastic activity in some lesions supported the presence of active remodeling in and around FFCD. Two femoral lesions in our series also revealed a dense fibrous tissue containing small areas of cartilaginous differentiation, which was confirmed by S-100 immunostaining. There was not any separate, discrete fibrous band in the vicinity of the lesion. However, we found firm fibrous tissue, similar to a periosteal anlage, tightly adhering to the overlying thickened periosteum in the notch of the cortex.

Resolution, persistence, or progression of the deformity

The compiled data (Table 1) revealed that the potential for spontaneous resolution of the lesion with resultant disappearance of the deformity appeared to vary depending on the affected site. In 37 of 46 cases, there was enough documented information to determine the clinical course of the lesion. There were 19 spontaneous resolutions or partial improvements, which were observed in 17 tibiae (1,3,4,7,8,10,11,13,19, and ours) and two ulnas (14, and ours). Deformity persisted in five tibiae (4,10,17, and ours) and one femur (ours), whereas progression of the deformity was observed in seven tibiae (3,13,16,18, and ours), four femora (1,15, and ours), and one humerus (14). There was no detailed documentation regarding the clinical course in nine of the tibial lesions (1,4,5,7,13,17,18). The compiled data indicated that at least 17 (45%) of the 38 tibial lesions and all of the ulnar lesions healed spontaneously. In contrast, all of the femoral and humeral lesions showed an increase or persistence of the deformity.

Treatment, follow-up, complications, and residual problems

Conservative treatment, by either observation only or bracing, was successful in 13 tibial and two ulnar FFCDs. Surgical interventions included biopsy only in four tibiae; hemicircumferential periosteal release in one femur; hemicircumferential periosteal release with incomplete osteotomy in one tibia; and corrective osteotomy in 18 tibiae, four femora, and one humerus, and nonspecified in one tibia. In general, the final outcomes in the surgically treated cases were satisfactory. Postoperative complications were associated with proximal tibial osteotomy: peroneal nerve palsy in two tibiae (1,4) and mild, residual genu valgum in two tibiae (4, and ours). Residual limb shortening in spite of resolution of the lesion was seen in one humerus (14) and two ulnas (14, and ours).

DISCUSSION

Regardless of the site of involvement, the radiographic features of FFCD on plain radiographs and magnetic resonance imaging (MRI) are essentially identical in all instances. Plain radiographs in affected tibiae demonstrate a well-defined focal area of lucency in the proximal medial cortex with sclerotic cortical thickening extending distally (1,3,5–7,13). In contrast, the radiographs of FFCD of the femur and ulna show similar focal radiolucent lesions in the distal cortex with cortical thickening extending proximally (1,14). MRI findings are low signal density on both short TR/short TE and long TR/long TE images, consistent with the dense fibroconnective tissue seen on pathologic examination. The sclerotic lesion, initially located at the metaphysis or metaphyseodiaphyseal junction, migrates into diaphysis with the longitudinal growth of the long bone.

Our observation of the histopathology of FFCD was in agreement with that of other authors (3,4,10,17–19), who have suggested that the lesions showed various histopathologic features, ranging from purely dense fibrous tendon-like tissue to benign fibrocartilaginous tissue. In our series, one of the three surgically treated tibial lesions (case 5) was composed purely of fibrous tissue. In one other case (case 6), the tibial lesion contained both hyaline and fibrocartilage, in addition to fibrous tissue. The remaining tibial and two femoral surgically treated cases (cases 1, 2, and 7) contained both fibrous and fibrocartilaginous elements. We observed that the presence of fibrocartilage was closely associated with the cellularity of the lesion. The association of fibroblasts and cartilage cells and the absence of cartilage in the hyalinized fibrous nodule strongly suggest that FFCD may undergo evolutionary changes from its initial cellular phase in cartilaginous form to its late paucicellular phase in a more fibrous form (12). Two femoral lesions were found to be histologically identical to their tibial counterparts, but lacked an obvious fibrous band in the vicinity, a characteristic of the cases in Beaty and Barrett's series (2). Previous authors (1,15) have also reported that fibrous bands were not observed in femoral FFCD.

We believe that the term FFCD may not be relevant and can potentially mislead people who are not familiar with this entity. First, the term fibrocartilaginous dysplasia has been used to describe another pathologic process of the long bone (fibrous dysplasia) in which cartilaginous differentiation can be conspicuous (9); second, the presence of fibrocartilage is not an essential feature; and third, age-related fibrocartilaginous metaplasia in tendons and ligaments is not uncommon. Because the lesion is fairly well circumscribed, closely related to the periosteum, and histologically composed of fibrous and/or cartilaginous tissue, the term "subperiosteal fibrocartilaginous pseudotumor of long bones" was proposed (12).

The natural history of FFCD is still being studied, and little is known about the exact etiologic and prognostic factors. Accordingly, treatment has been optional. Our investigation indicates that at least 45% of the cases of tibial FFCD showed spontaneous resolution or progressive improvement of the deformity. We believe that the proportion of cases that may not need definitive treatment may be larger than previously documented, because many of the treated cases may have resolved on their own had they been given the opportunity of observation

without jumping in to surgical correction. Therefore, we agree with other authors that, when possible, the primary conservative approach is justified (3,4,10,13,18). However, excellent results can be obtained after proximal tibial osteotomy with or without resection of the lesion with little chance of recurrence (4,7,17,18). Complications after corrective osteotomy of proximal tibia, including postoperative valgus deformity and neurologic injury (peroneal nerve palsy) have been reported (1,4).

FFCD occurs very rarely in other long bones. So far, to the best of our knowledge, only five femoral, two ulnar, and one humeral FFCD have been documented. It was intriguing to notice that spontaneous resolution was observed in two ulnar FFCDs, but in none of the femoral or humeral FFCDs. These caused an increase or persistence of the deformity and were eventually treated with surgical correction. It is still unknown whether the natural healing course of FFCD of the long bone can vary depending on the affected site. Considering the identical histopathologic features of the femoral and humeral FFCDs to the tibial counterparts, there may be some possibility of spontaneous resolution in these cases. This question will be answered only when a larger series of FFCD cases occurring in other long bones is investigated.

We conclude that although FFCD of the long bone is a benign lesion, its histopathology and clinical course are not uniform, and thus regular clinical and radiologic examinations are essential. When the deformity increases or persists in spite of a reasonable period of observation or bracing, or when the deformity is severe enough to jeopardize adjacent joint mechanics and alignment, timely corrective osteotomy is then indicated.

REFERENCES

1. Albinana J, Cuervo M, Certucha JA, Gonzalez-Mediero I, Abril JC. Five additional cases of local fibrocartilaginous dysplasia. *J Pediatr Orthop Part B* 1997;6:52–5.
2. Beaty JH, Barrett IR. Unilateral angular deformity of the distal end of the femur secondary to a focal fibrous tether: a report of four cases. *J Bone Joint Surg Am* 1989;71:440–5.
3. Bell SN, Campbell PE, Cole WG, Menelaus MB. Tibia vara caused by focal fibrocartilaginous dysplasia: three cases report. *J Bone Joint Surg Br* 1985;67:780–4.
4. Bradish CF, Davies SJ, Malone M. Tibia vara due to focal fibrocartilaginous dysplasia: the natural history. *J Bone Joint Surg Br* 1988;70:106–8.
5. Cockshott WP, Martin R, Friedman L, Yuen M. Focal fibrocartilaginous dysplasia and tibia vara: a case report. *Skeletal Radiol* 1994;23:333–5.
6. Haasbeek JF, Rang MC, Blackburn N. Periosteal tether causing angular deformity: report of two clinical cases and an experimental model. *J Pediatr Orthop* 1995;15:677–81.
7. Herman TE, Siegel MJ, McAlister WH. Focal fibrocartilaginous dysplasia associated with tibia vara. *Radiology* 1990;177:767–8.
8. Husien AM, Kale VR. Tibia vara caused by focal fibrocartilaginous dysplasia. *Clin Radiol* 1989;40:104–5.
9. Ishida T, Dorfman HD. Massive chondroid differentiation in fibrous dysplasia of bone (fibrocartilaginous dysplasia). *Am J Surg Pathol* 1993;17:924–30.
10. Jouve JL, Debelenet H, Petit P, Guillaume JM, Gaudeuille A, Bollini G. Focal fibrocartilaginous dysplasia and tibia vara: apropos of 2 cases: review of the literature. *Rev Chir Orthop Reparatrice Appar Mot* 1997;84:473–6.
11. Kariya Y, Taniguchi K, Yagisawa H, Ooi Y. Focal fibrocartilaginous dysplasia: consideration of healing process. *J Pediatr Orthop* 1991;11:545–7.
12. Kim CJ, Choi IH, Cho TJ, Chung CY, Chi JG. The histopathologic spectrum of subperiosteal fibrocartilaginous pseudotumor of long bone (focal fibrocartilaginous dysplasia). *Pathol Int* 1999;49:1000–6.
13. Landreau-Jolivet I, Pilliard D, Taussig G. Unilateral tibia vara in young children caused by focal fibrocartilaginous dysplasia: apropos of 4 new cases. *Rev Chir Orthop Reparatrice Appar Mot* 1992;78:411–4.
14. Lincoln TL, Birch JG. Focal fibrocartilaginous dysplasia in the upper extremity. *J Pediatr Orthop* 1997;17:528–32.
15. Macnicol M. Focal fibrocartilaginous dysplasia of the femur. *J Pediatr Orthop Part B* 1999;8:61–3.
16. Marchiodi L, Stilli S, Di Gennaro G. Tibia vara caused by focal fibrocartilaginous dysplasia. *Chir Organi Mov* 1995;80:453–6.
17. Meyer JS, Davidson RS, Hubbard AM, Conard KA. MRI of focal fibrocartilaginous dysplasia. *J Pediatr Orthop* 1995;15:304–6.
18. Olney BW, Cole WG, Menelaus MB. Three additional cases of focal fibrocartilaginous dysplasia causing tibia vara. *J Pediatr Orthop* 1990;10:405–7.
19. Zayer M. Tibia vara in focal fibrocartilaginous dysplasia: a report of 2 cases. *Acta Orthop Scand* 1992;63:353–5.